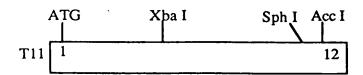
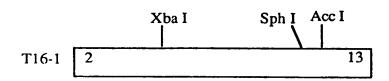
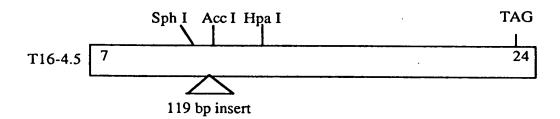
FIGURE 1 PARTIAL cDNA CLONES OF THE CFTR GENE







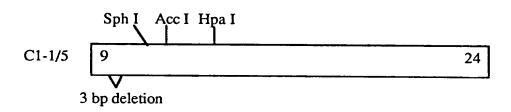




FIGURE 2
STRATEGY FOR CONSTRUCTING pKK- CFTR1

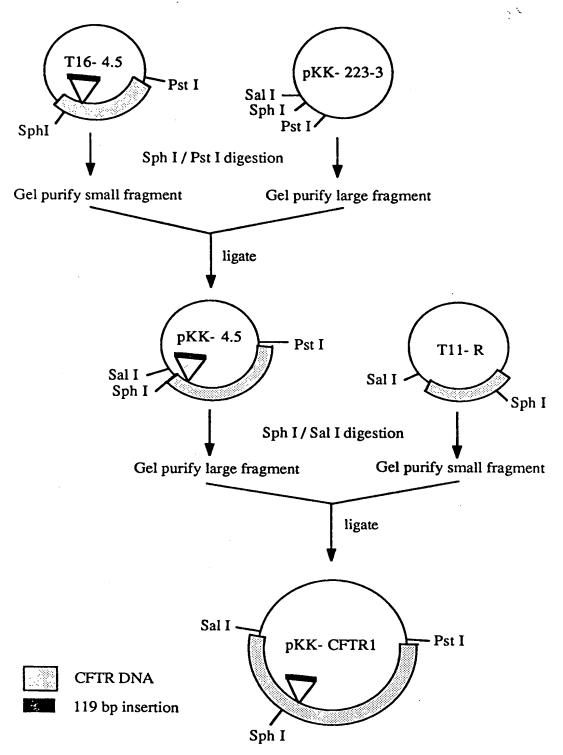


FIGURE 3
CONSTRUCTION OF THE pKK- CFTR2 PLASMID

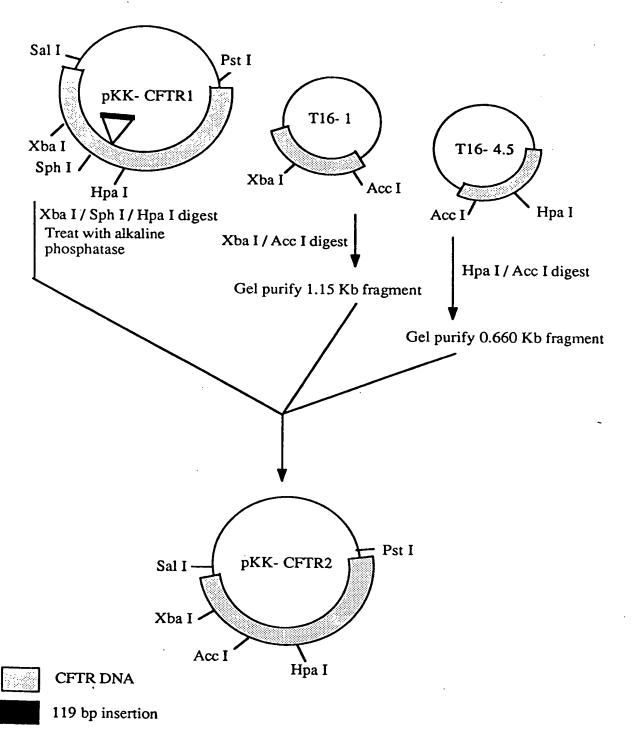


FIGURE 4
STRATEGY FOR CONSTRUCTING THE pSC- CFTR2 PLASMID

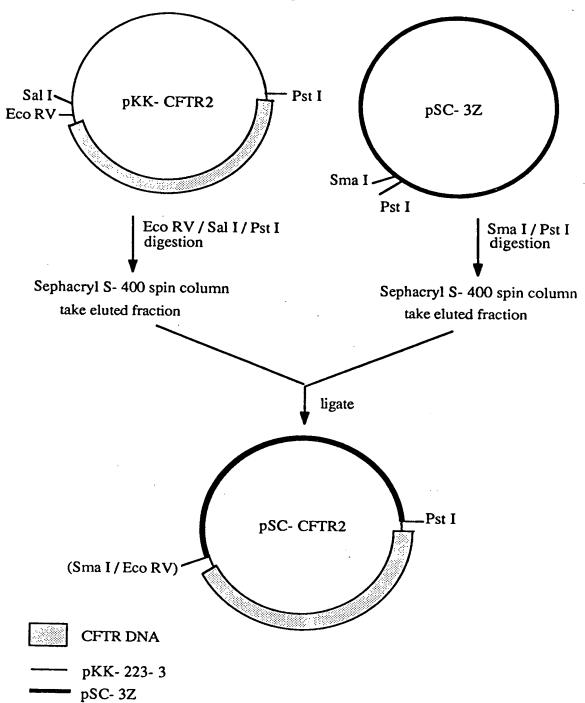
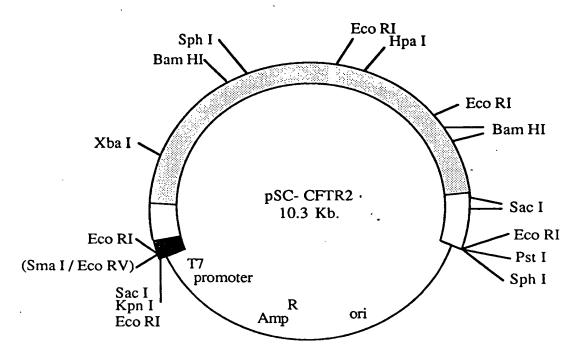


FIGURE 5
MAP OF pSC- CFTR2



CFTR coding region

CFTR noncoding region

T11- derived non- CFTR DNA

pSC- 3Z

p	CTGAGGTGACAATGACATCTACTCTGACATTCTCTCCTCAGGACATCTCCAAGTTTGCAG GACTCCACTGTTACTGTAGAGACTGTAAGAGGAGTCCTGTAGAGGTTCAAACGTC
---	--

FIGURE 7A

CONSTRUCTION OF THE pKK- CFTR3 cDNA

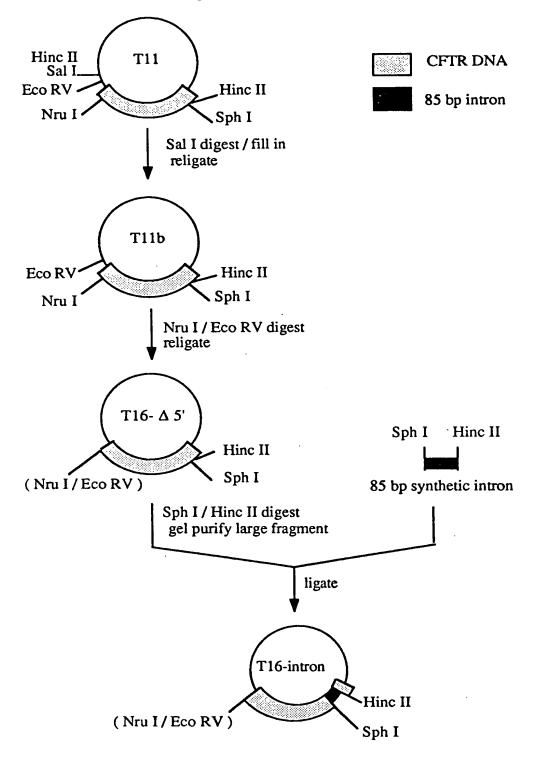


FIGURE 7B

CONSTRUCTION OF THE pKK- CFTR3 CLONE (cont'd.)

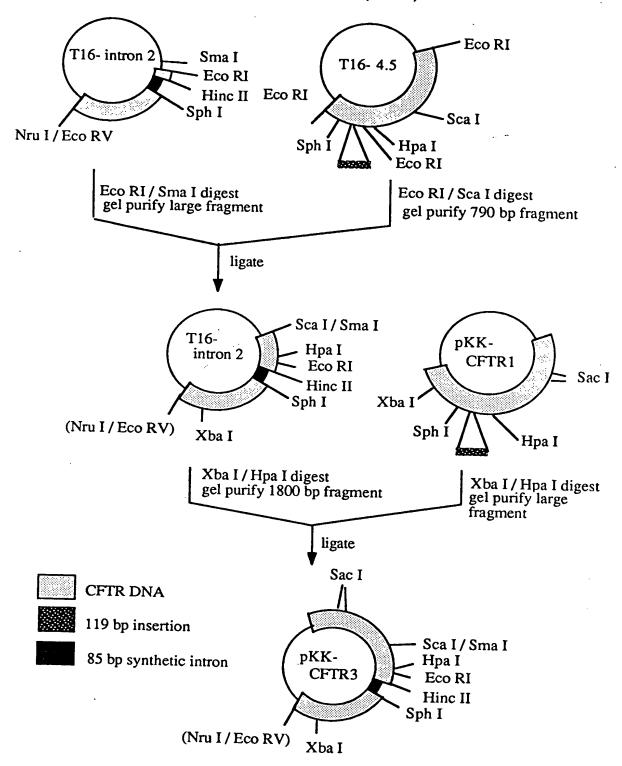
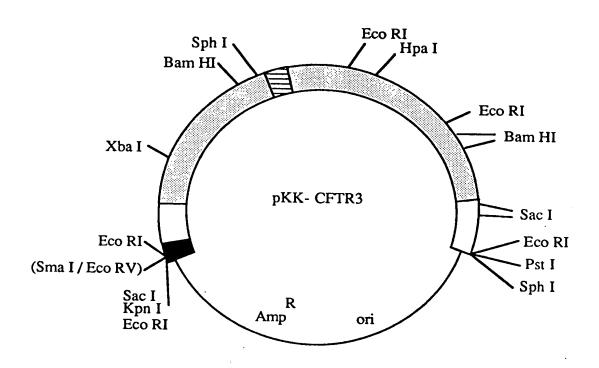


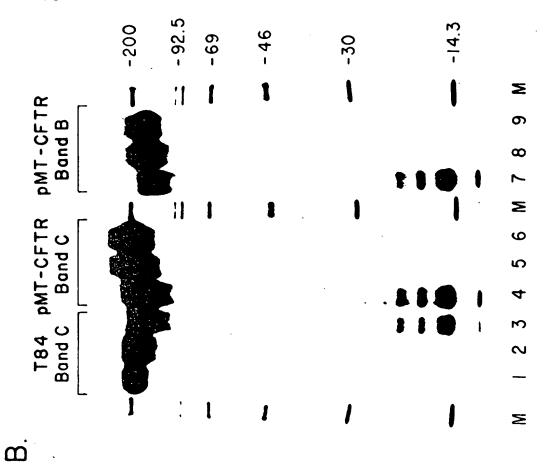
FIGURE 8
MAP OF pKK- CFTR3

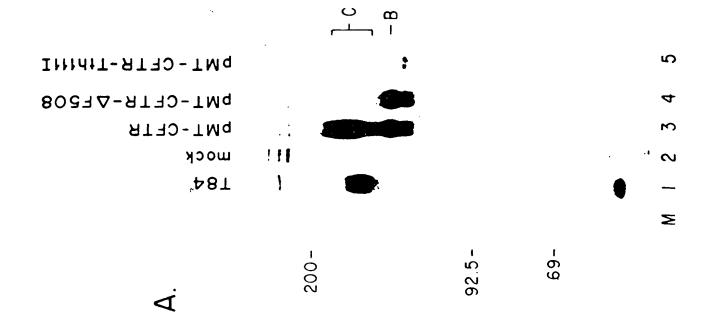


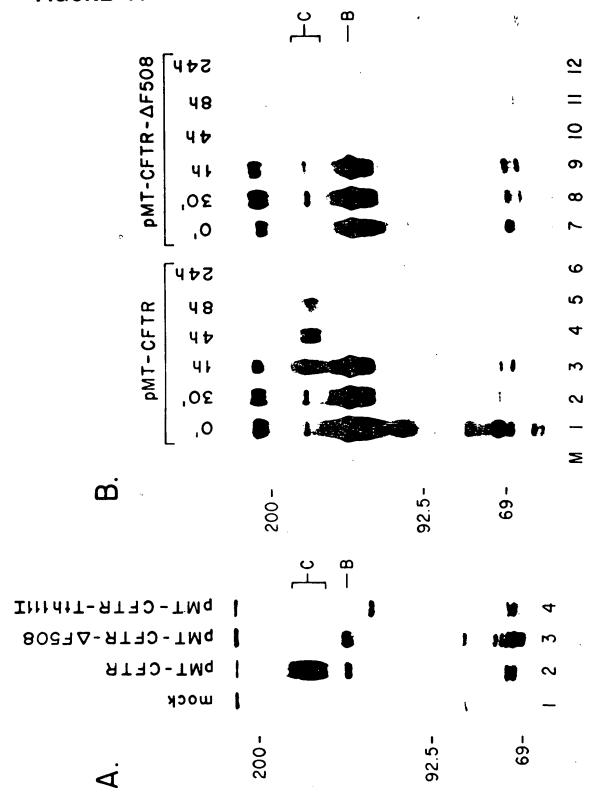
: - **;	CFTR coding region
	CFTR noncoding region
	85 bp intron
	T11- derived non- CFTR DNA
	pKK- 223- 3

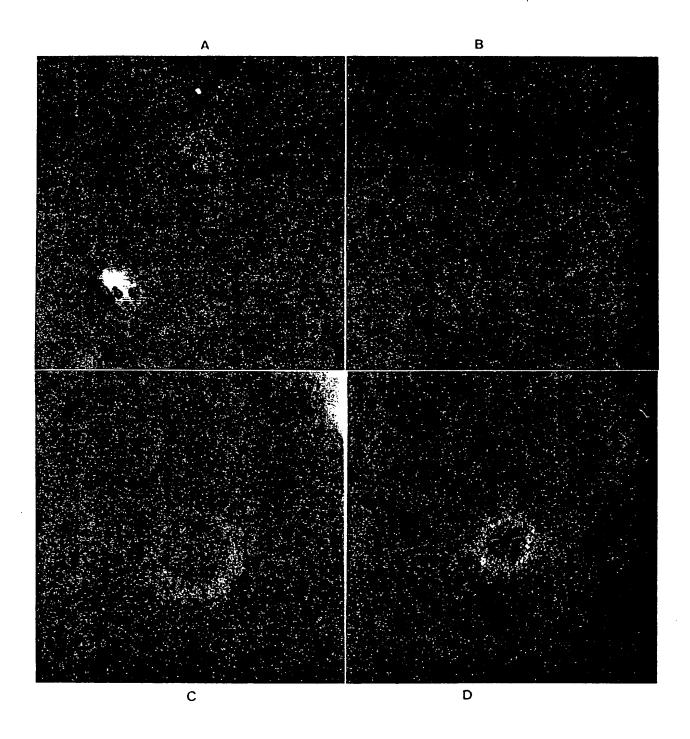
IG 4-9.2

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IG 4-9.2

pMT-CFTR-K464M
pMT-CFTR-K1250M
pMT-CFTR-A1507
pMT-CFTR-deglycos.
pMT-CFTR-R334W

200-



92.5-

IG 4-9.2

PABLE 1 Figure 15

1	AATTGGAAGCAAATGACATCACAGCAGGTCAGAGAAAAAGGG	42
43	TTGAGCGGCAGCCACCCAGAGTAGTAGGTCTTTGGCATTAGG	84
85	AGCTTGAGCCCAGACGCCCTAGCAGGGACCCCAGCGCCCGA	126
1	MetGlnArgSerProLeuGluLysAlaSerValVal	12
127	GAGACCATGCAGAGGTCGCCTCTGGAAAAGGCCAGCGTTGTC	168
13	SerLysLeuPhePheSerTrpThrArgProIleLeuArgLys	26
169	TCCAAACTTTTTTCAGCTGGACCAGACCAATTTTGAGGAAA	210
27	GlyTyrArgGlnArgLeuGluLeuSerAspIleTyrGlnIle	40
211	GGATACAGACAGCGCCTGGAATTGTCAGACATATACCAAATC	252
41	ProSerValAspSerAlaAspAsnLeuSerGluLysLeuGlu	54
253	CCTTCTGTTGATTCTGCTGACAATCTATCTGAAAAATTGGAA	294
55	ArgGluTrpAspArgGluLeuAlaSerLysLysAsnProLys	68
295	AGAGAATGGGATAGAGAGCTGGCTTCAAAGAAAAATCCTAAA	336
69	LeuIleAsnAlaLeuArgArgCysPhePheTrpArgPheMet	82
337	CTCATTAATGCCCTTCGGCGATGTTTTTTCTGGAGATTTATG	378
83	PheTyrGlyIlePheLeuTyrLeuGlyGluValThrLysAla	96
379	TTCTATGGAATCTTTTATATTTAGGGGAAGTCACCAAAGCA	420
97	ValGlnProLeuLeuGlyArgIleIleAlaSerTyrAsp	110
421	GTACAGCCTCTCTTACTGGGAAGAATCATAGCTTCCTATGAC	462
111	ProAspAsnLysGluGluArgSerIleAlaIleTyrLeuGly	124
463	CCGGATAACAAGGAGGAACGCTCTATCGCGATTTATCTAGGC	504
125	IleGlyLeuCysLeuLeuPheIleValArgThrLeuLeuLeu	138
505	ATAGGCTTATGCCTTCTCTTTATTGTGAGGACACTGCTCCTA	546
139	HisProAlaIlePheGlyLeuHisHisIleGlyMetGlnMet	152
547	CACCCAGCCATTTTTGGCCTTCATCACATTGGAATGCAGATG	588
153	ArgIleAlaMetPheSerLeuIleTyrLysLysThrLeuLys	166
589	AGAATAGCTATGTTTAGTTTGATTTATAAGAAGACTTTAAAG	630
167	LeuSerSerArgValLeuAspLysIleSerIleGlyGlnLeu	180
631	CTGTCAAGCCGTGTTCTAGATAAAATAAGTATTGGACAACTT	672
181	ValSerLeuLeuSerAsnAsnLeuAsnLysPheAspGluGly	194
673	GTTAGTCTCCTTTCCAACAACCTGAACAAATTTGATGAAGGA	714

. 195	LeuAlaLeuAlaHisPheValTrpIleAlaProLeuGlnVal	208
715	CTTGCATTGGCACATTTCGTGTGGATCGCTCCTTTGCAAGTG	756
· 209 757	AlaLeuLeuMetGlyLeuIleTrpGluLeuLeuGlnAlaSer GCACTCCTCATGGGGCTAATCTGGGAGTTGTTACAGGCGTCT	222 798
223	AlaPheCysGlyLeuGlyPheLeuIleValLeuAlaLeuPhe	236
799	GCCTTCTGTGGACTTGGTTTCCTGATAGTCCTTGCCCTTTTT	840
237	GlnAlaGlyLeuGlyArgMetMetMetLysTyrArgAspGln	250
841	CAGGCTGGGCTAGGGAGAATGATGAAGTACAGAGATCAG	882
251	ArgAlaGlyLysIleSerGluArgLeuValIleThrSerGlu	264
883	AGAGCTGGGAAGATCAGTGAAAGACTTGTGATTACCTCAGAA	924
265	MetIleGluAsnIleGlnSerValLysAlaTyrCysTrpGlu	278
925	ATGATTGAAAATATCCAATCTGTTAAGGCATACTGCTGGGAA	966
279	GluAlaMetGluLysMetIleGluAsnLeuArgGlnThrGlu	292
967	GAAGCAATGGAAAAAATGATTGAAAACTTAAGACAAACAGAA	1008
293	LeuLysLeuThrArgLysAlaAlaTyrValArgTyrPheAsn	306
1009	CTGAAACTGACTCGGAAGGCAGCCTATGTGAGATACTTCAAT	1050
307	SerSerAlaPhePhePheSerGlyPhePheValValPheLeu	320
1051	AGCTCAGCCTTCTTCTCAGGGTTCTTTGTGGTGTTTTTA	1092
321 1093	SerValLeuProTyrAlaLeuIleLysGlyIleIleLeuArg TCTGTGCTTCCCTATGCACTAATCAAAGGAATCATCCTCCGG	334 1134
335 1135		348 1176
349	AlaValThrArgGlnPheProTrpAlaValGlnThrTrpTyr	362
1177	GCGGTCACTCGGCAATTTCCCTGGGCTGTACAAACATGGTAT	1218
363	AspSerLeuGlyAlaIleAsnLysIleGlnAspPheLeuGln	376
1219	GACTCTCTTGGAGCAATAAACAAAATACAGGATTTCTTACAA	1260
377	LysGlnGluTyrLysThrLeuGluTyrAsnLeuThrThrThr	390
1261	AAGCAAGAATATAAGACATTGGAATATAACTTAACGACTACA	1302
391	GluValValMetGluAsnValThrAlaPheTrpGluGluGly	404 ⁻
1303	GAAGTAGTGATGGAGAATGTAACAGCCTTCTGGGAGGAGGGA	1344
405	PheGlyGluLeuPheGluLysAlaLysGlnAsnAsnAsnAsn	418
1345	TTTGGGGAATTATTTGAGAAAGCAAAACAAAACAATAACAAT	1386

.

•	419	ArgLysThrSerAsnGlyAspAspSerLeuPhePheSerAsn	432
	1387	AGAAAAACTTCTAATGGTGATGACAGCCTCTTCTTCAGTAAT	1428
	1507	NOMANACTICIANTOGIGATOACAGCCICITCTICAGIAMI	1426
	433	DhoCowlouI auClamba Da-11-11 auta-4 a 71 a mi	
		PheSerLeuLeuGlyThrProValLeuLysAspIleAsnPhe	446
	1429	TTCTCACTTCTTGGTACTCCTGTCCTGAAAGATATTAATTTC	1470
	,,,		
	447	LysIleGluArgGlyGlnLeuLeuAlaValAlaGlySerThr	460
	1471	AAGATAGAAAGAGGACAGTTGTTGGCGGTTGCTGGATCCACT	1512
•			
	461	GlyAlaGlyLysThrSerLeuLeuMetMetIleMetGlyGlu	474
	1513	GGAGCAGGCAAGACTTCACTTCTAATGATGATTATGGGAGAA	1554
•		·	
	475	LeuGluProSerGluGlyLysIleLysHisSerGlyArgIle	488
	1555	CTGGAGCCTTCAGAGGGTAAAATTAAGCACAGTGGAAGAATT	1596
	489	SerPheCysSerGlnPheSerTrpIleMetProGlyThrIle	502
	1597	TCATTCTGTTCTCAGTTTTCCTGGATTATGCCTGGCACCATT	1638
	503	LysGluAsnIleIlePheGlyValSerTyrAspGluTyrArg	516
	1639	AAAGAAAATATCATCTTTGGTGTTTCCTATGATGAATATAGA	1680
			1000
	517	TyrArgSerValIleLysAlaCysGlnLeuGluGluAspIle	530
	1681	TACAGAAGCGTCATCAAAGCATGCCAACTAGAAGAGGACATC	1722
•			1/22
	531	SerLysPheAlaGluLysAspAsnIleValLeuGlyGluGly	544
	1723	TCCAAGTTTGCAGAGAAAGACAATATAGTTCTTGGAGAAGGT	1764
		TOTAL	1704
	545	GlyIleThrLeuSerGlyGlyGlnArgAlaArgIleSerLeu	558
	1765	GGAATCACACTGAGTGGAGGTCAACGAGCAAGAATTTCTTTA	1806
		COLUMN TO THE CONTROL OF THE CONTROL OF THE COLUMN TO THE	1606
	559	AlaArgAlaValTyrLysAspAlaAspLeuTyrLeuLeuAsp	570
	1807	GCAAGAGCAGTATACAAAGATGCTGATTTGTATTTATTAGAC	572
	1007	COMMONGCAGINIACAMAGNIGCIGNIIIGINIIINIINGAC	1848
	573	SerProPhoCluTuriouAceNelloumbuclutu-clutu	504
	1849	SerProPheGlyTyrLeuAspValLeuThrGluLysGluIle TCTCCTTTTGGATACCTAGATGTTTTAACAGAAAAAGAAATA	586
	1047	TOTOCTTTTGGATACCTAGATGTTTTAACAGAAAAAGAAATA	1890
	587	PhoCluCorCyclic Control and and and an and	
	1891	PheGluSerCysValCysLysLeuMetAlaAsnLysThrArg	600
	1091	TTTGAAAGCTGTGTCTGTAAACTGATGGCTAACAAAACTAGG	1932
	601	The Louve The Continuous Charter to the Cont	-
	1933	IleLeuValThrSerLysMetGluHisLeuLysLysAlaAsp	614
•	1733	ATTTTGGTCACTTCTAAAATGGAACATTTAAAGAAAGCTGAC	1974
	615	Ivellel outlet outlet outlet or on the	
	1975	LysIleLeuIleLeuHisGluGlySerSerTyrPheTyrGly	628
	17/3	AAAATATTAATTTTGCATGAAGGTAGCAGCTATTTTTATGGG	2016
	620	Thunha Carrolla Language	
	629 2017	ThrPheSerGluLeuGlnAsnLeuGlnProAspPheSerSer	642
	2017	ACATTTTCAGAACTCCAAAATCTACAGCCAGACTTTAGCTCA	2058

643	LysLeuMetGlyCysAspSerPheAspGlnPheSerAlaGlu	656
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657	ArgArgAsnSerIleLeuThrGluThrLeuHisArgPheSer	670
2101	AGAAGAAATTCAATCCTAACTGAGACCTTACACCGTTTCTCA	2142
671	LeuGluGlyAspAlaProValSerTrpThrGluThrLysLys	684
2143	TTAGAAGGAGATGCTCCTGTCTCCTGGACAGAAACAAAAAA	2184
685	GlnSerPheLysGlnThrGlyGluPheGlyGluLysArgLys	698
2185	CAATCTTTTAAACAGACTGGAGAGTTTGGGGAAAAAAGGAAG	2226
699	AsnSerIleLeuAsnProIleAsnSerIleArgLysPheSer	712
2227	AATTCTATTCTCAATCCAATCAACTCTATACGAAAATTTTCC	2268
713	IleValGlnLysThrProLeuGlnMetAsnGlyIleGluGlu	726
2269	ATTGTGCAAAAGACTCCCTTACAAATGAATGGCATCGAAGAG	2310
727	AspSerAspGluProLeuGluArgArgLeuSerLeuValPro	740
2311	GATTCTGATGAGCCTTTAGAGAGAGGCTGTCCTTAGTACCA	2352
741	AspSerGluGlnGlyGluAlaIleLeuProArgIleSerVal	754
2353	GATTCTGAGCAGGGAGAGGCGATACTGCCTCGCATCAGCGTG	2394
755	IleSerThrGlyProThrLeuGlnAlaArgArgArgGlnSer	768
2395	ATCAGCACTGGCCCCACGCTTCAGGCACGAAGGAGGCAGTCT	2436
769	ValLeuAsnLeuMetThrHisSerValAsnGlnGlyGlnAsn	782
2437	GTCCTGAACCTGATGACACTCAGTTAACCAAGGTCAGAAC	2478
783	IleHisArgLysThrThrAlaSerThrArgLysValSerLeu	796
2479	ATTCACCGAAAGACAACAGCATCCACACGAAAAGTGTCACTG	2520
797	AlaProGlnAlaAsnLeuThrGluLeuAspIleTyrSerArg	810
2521	GCCCCTCAGGCAAACTTGACTGAACTGGATATATTCAAGA	2562
811	ArgLeuSerGlnGluThrGlyLeuGluIleSerGluGluIle	824
2563	AGGTTATCTCAAGAAACTGGCTTGGAAATAAGTGAAGAAATT	2604
825	AsnGluGluAspLeuLysGluCysLeuPheAspAspMetGlu	838
2605	AACGAAGAAGTTAAAGGAGTGCCTTTTTGATGATATGGAG	2646
839	SerIleProAlaValThrThrTrpAsnThrTyrLeuArgTyr	852
2647	AGCATACCAGCAGTGACTACATGGAACACATACCTTCGATAT	2688
853	IleThrValHisLysSerLeuIlePheValLeuIleTrpCys	866
2689	ATTACTGTCCACAAGAGCTTAATTTTTGTGCTAATTTGGTGC	2730

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867	LeuValllePheLeuAlaGluValAlaAlaSerLeuValVal	880
2731	TTAGTAATTTTTCTGGCAGAGGTGGCTGCTTCTTTGGTTGTG	2772
881	LeuTrpLeuLeuGlyAsnThrProLeuGlnAspLysGlyAsn	894
2773	CTGTGGCTCCTTGGAAACACTCCTCTTCAAGACAAAGGGAAT	2814
895	SerThrHisSerArgAsnAsnSerTyrAlaVallleIleThr	908
2815	AGTACTCATAGTAGAAATAACAGCTATGCAGTGATTATCACC	2856
909	SerThrSerSerTyrTyrValPheTyrIleTyrValGlyVal	922
2857	AGCACCAGTTCGTATTATGTGTTTTACATTTACGTGGGAGTA	2898
923	AlaAspThrLeuLeuAlaMetGlyPhePheArgGlyLeuPro	936
2899	GCCGACACTTTGCTTGCTATGGGATTCTTCAGAGGTCTACCA	2940
937	LeuValHisThrLeuIleThrValSerLysIleLeuHisHis	950
2941	CTGGTGCATACTCTAATCACAGTGTCGAAAATTTTACACCAC	2982
951	LysMetLeuHisSerValLeuGlnAlaProMetSerThrLeu	964
2983	AAAATGTTACATTCTGTTCTTCAAGCACCTATGTCAACCCTC	3024
965	AsnThrLeuLysAlaGlyGlyIleLeuAsnArgPheSerLys	978
3025	AACACGTTGAAAGCAGGTGGGATTCTTAATAGATTCTCCAAA	3066
979	AspIleAlaIleLeuAspAspLeuLeuProLeuThrIlePhe	992
3067	GATATAGCAATTTTGGATGACCTTCTGCCTCTTACCATATTT	3108
993 3109	AspPheIleGlnLeuLeuLeuIleValIleGlyAlaIleAla GACTTCATCCAGTTGTTATTAATTGTGATTGGAGCTATAGCA	1006 3150
1007	ValValAlaValLeuGlnProTyrIlePheValAlaThrVal	1020
3151	GTTGTCGCAGTTTTACAACCCTACATCTTTGTTGCAACAGTG	3192
1021	ProVallleValAlaPhelleMetLeuArgAlaTyrPheLeu	1034
3193	CCAGTGATAGTGGCTTTTATTATGTTGAGAGCATATTTCCTC	3234
1035	GlnThrSerGlnGlnLeuLysGlnLeuGluSerGluGlyArg	1048
3235	CAAACCTCACAGCAACTCAAACAACTGGAATCTGAAGGCAGG	3276
1049	SerProIlePheThrHisLeuValThrSerLeuLysGlyLeu	1062
3277	AGTCCAATTTTCACTCATCTTGTTACAAGCTTAAAAGGACTA	3318
1063	TrpThrLeuArgAlaPheGlyArgGlnProTyrPheGluThr	1076
3319	TGGACACTTCGTGCCTTCGGACGCCAGCCTTACTTTGAAACT	3360
1077	LeuPheHisLysAlaLeuAsnLeuHisThrAlaAsnTrpPhe	1090
3361	CTGTTCCACAAAGCTCTGAATTTACATACTGCCAACTGGTTC	3402

·

1091 3403		1104 3444
1105		1118
3445	ATGATTTTTGTCATCTTCTTCATTGCTGTTACCTTCATTTCC	3486
1119 3487		1132 3528
1133		1146
3529		3570
1147 3571		1160 3612
1161 3613		1174
		3654
1175 3655		1188 3696
1189 3697		1202 3738
1203		1216
3739		3780
1217 3781		1230
		3822
1231 3823		1244 3864
1245		1258
3865		3906
1259		1272
3907	AGACTACTGAACACTGAAGGAGAAATCCAGATCGATGGTGTG	3948
1273		1286
3949	TCTTGGGATTCAATAACTTTGCAACAGTGGAGGAAAGCCTTT	3990
1287		1300
3991	GGAGTGATACCACAGAAAGTATTTATTTTTTTCTGGAACATTT	4032
1301	ArgLysAsnLeuAspProTyrGluGlnTrpSerAspGlnGlu	1314
4033		4074

.

	1315	IleTrpLysValAlaAspGluValGlyLeuArgSerValIle	1328
	4075	ATATGGAAAGTTGCAGATGAGGTTGGGCTCAGATCTGTGATA	4116
	4075	NINIOOMMIO 1000000000000000000000000000000000000	
•	1329	GluGlnPheProGlyLysLeuAspPheValLeuValAspGly	1342
		GAACAGTTTCCTGGGAAGCTTGACTTTGTCCTTGTGGATGGG	4158
	4117	GAACAGIIICCIGGGAAGCIIGACIIITTTTTTTTTTTTT	
		GlyCysValLeuSerHisGlyHisLysGlnLeuMetCysLeu	1356
	1343	GGCTGTGTCCTAAGCCATGGCCACAAGCAGTTGATGTGCTTG	4200
_	4159	GGCTGTGTCCTAAGCCATGGCCACAAGCAGTTGATGTGCTTG	1.200
•		and the state of t	1370
	1357	AlaArgSerValLeuSerLysAlaLysIleLeuLeuLeuAsp	4242
	4201	GCTAGATCTGTTCTCAGTAAGGCGAAGATCTTGCTGCTTGAT	4242
•			120/
	1371	GluProSerAlaHisLeuAspProValThrTyrGlnIleIle	1384
	4243	GAACCCAGTGCTCATTTGGATCCAGTAACATACCAAATAATT	4284
	1385	ArgArgThrLeuLysGlnAlaPheAlaAspCysThrValIle	1398
	4285	AGAAGAACTCTAAAACAAGCATTTGCTGATTGCACAGTAATT	4326
	7203	1011101110111011	
	1399	LeuCysGluHisArgIleGluAlaMetLeuGluCysGlnGln	1412
	4327	CTCTGTGAACACAGGATAGAAGCAATGCTGGAATGCCAACAA	4368
	4321	Cicididanonomonimonario	
	1413	PheLeuValIleGluGluAsnLysValArgGlnTyrAspSer	1426
		TTTTTGGTCATAGAAGAGAACAAAGTGCGGCAGTACGATTCC	4410
	4369	TTTTTGTCATAGAAGAAGAAGAAGTGCGGGGGTGCTTCGTTTTT	
	1/07	IleGlnLysLeuLeuAsnGluArgSerLeuPheArgGlnAla	1440
	1427	ATCCAGAAACTGCTGAACGAGAGGAGCCTCTTCCGGCAAGCC	4452
	4411	ATCCAGAAACIGCIGAACGAGAGGAGCCICIICCGGCAAGCC	4.132
		The Declination of the Declination	1454
	1441	IleSerProSerAspArgValLysLeuPheProHisArgAsn	4494
	4453	ATCAGCCCTCCGACAGGGTGAAGCTCTTTCCCCACCGGAAC	4474
			1468
	1455	SerSerLysCysLysSerLysProGlnIleAlaAlaLeuLys	4536
	4495	TCAAGCAAGTGCAAGTCTAAGCCCCAGATTGCTGCTCTGAAA	4536
			4 4 0 0
	1469	GluGluThrGluGluGluValGlnAspThrArgLeuEnd	1482
	4537	GAGGAGACAGAAGAGAGGTGCAAGATACAAGGCTTTAGAGA	4578
	4579	GCAGCATAAATGTTGACATGGGACATTTGCTCATGGAATTGG	4620
	4621	AGCTCGTGGGACAGTCACCTCATGGAATTGGAGCTCGTGGAA	4662
	4663		4704
	4705		4746
	4747		4788
	4747		4830
	4831		4872
	4873	INNIIOTOTOTOTOTOTOTOTOTOTOTOTOTOTOTOTOTO	

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